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Postoperative fatal hypothermia in hydranencephaly with pre-operative hypothermia and a nerve palsy: A case report and review of the literature

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Abstract

Hydranencephaly is a rare condition characterised by complete or near complete absence of the cerebral hemispheres within relatively normal sized meninges and skull, the resulting cavity being filled with cerebrospinal fluid.

The following is a case report of a five month old hydranencephalic child with right upper motor facial nerve palsy who presented with signs of hydrocephalus who developed intractable hypothermia rapidly post ventriculo-peritoneal shunt insertion and demised. Her preoperative condition was associated with hypothermia.

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Introduction

Hydranencephaly is a rare condition characterised by complete or near complete absence of the cerebral hemispheres within relatively normal sized meninges and skull, the resulting cavity being filled with cerebrospinal fluid.[1,2,4] It is considered to be an extreme form of porencephaly, a rare disorder characterised by a cyst or a cavity in the cerebral hemispheres. Some authors have referred to this disorder as schizencephaly.[2]

The word hydranencephaly is a fusion of the words hydrocephalus and anencephaly as named by Crueilhier though this is in itself a distinct disorder.[2, 3] Hydranencephaly is a rare isolated abnormality occurring in less than 1 per 10 000 births worldwide.[3,10] About 1% of infants thought to have hydrocephalus clinically are later found to have hydranencephaly.[18] It occurs in all races without any sex predilection.[3] The life expectancy of most patients with hydranencephaly is markedly reduced, many being either stillborn or surviving less than a year. Documented causes of death

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for those born alive include dehydration, malnutrition and infections. Survival of up to 19years has been documented in the literature with some postulating that a survival of up to 30years can be obtained with aggressive nursing care.[3,5]

The following is a case report of a five month old hydranencephalic child who presented with signs of hydrocephalus. Considering that some of the children with this malformation survive for a long time, insertion of a ventriculo-peritoneal shunt is considered prudent to make child care easier.

Case Report

A five months old female infant was referred from a rural district hospital with a three months history of an abnormally enlarging head, blindness and a bulging anterior fontanel.

The mother had been booked and foetal ultrasound scan was not done during pregnancy. No maternal illnesses were experienced in pregnancy. She delivered the baby prematurely at 30 weeks of gestation.

The child had neonatal jaundice that lasted three weeks. Treatment was by sun exposure until the jaundice had cleared. She was admitted and treated of bronchopneumonia and a thigh abscess at two months of age. This is the time when the child was noted to be blind.

Developmentally, she started smiling at two months of age. She was not able to sit at five months and the parents were suspecting that this was because of the enlarged head.

The child was breastfeeding normally. She had now been started on weaning feeds (porridge) and was tolerating it very well. This was a well nourished child.

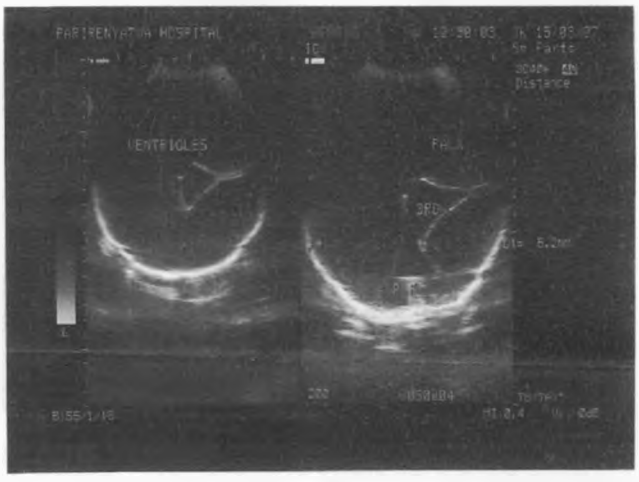
She was lagging behind on immunizations, having missed two doses each of the DPT (Diptheria, Pertussis and Tetanus) and HBV (Hepatitis B) vaccines.

This child was staying with both parents in the rural areas. There was no family history of congenital birth defects. The child was born as the first child in a first pregnancy for the non-consanguineously married couple.

On examination, the child was noted to be an alert child with an obviously enlarged head, which was brachycephalic. Head circumference was 45cm(normal expected range for age is 37,3-41,7cm).[12] She had low set ears, hypertelorism and was noted to be blind. She had a tense bulging fontanel with separation of the sagittal and coronal sutures. She had enlarged scalp vessels. A right seventh nerve upper motor neuron palsy was also noted. She had normal heart sounds and a pulse of 128beats per minute. Respiratory and abdominal examinations were normal. She had normal female genitalia.

An ultrasound scan was done which showed a normal posterior fossa and very little supratentorial tissue (figure 1).

Figure 1: Ultrasound scan picture for the infant's head.



Results for the full blood count were as follows: White cell count-10,2*10^9 /l Hemoglobin-10,23g/dl Platelets 393000/ul and Mean Corpuscular Volume 75,3fl. The blood chemistry was as follows: Na^+ 155mmol/l K^+ 4,6mmol/l Urea 4,6mmol/l and creatinine 39umol/l and random blood sugar of 6,0mmol/l.

A decision for insertion of a palliative shunt was made to arrest further head enlargement and to ease up martenal care. The child had episodes of hypothermia which were later noted in retrospect with temperatures between 34-36°C. She was starved for six hours pre-operatively. The child was noted to be having a very weak cry on the operating table.

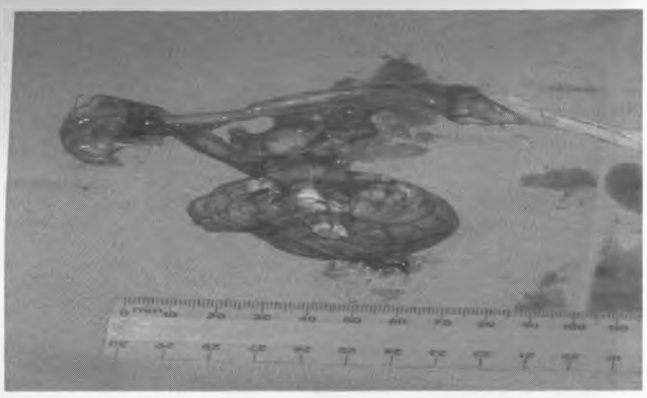
A ventriculo-peritoneal shunt of the Harare shunt type was inserted with a warming mattress and wool cover being used for heat conservation. At completion of the operation the child was hypothermic and remained so despite all attempts to warm her up. About six hours post-operatively, the child had a cardiorespiratory arrest and resuscitation attempts were unsuccessful.

The post-mortem conducted revealed congenital absence of both cerebral hemispheres. The diencephalon was however present. The tentorium and falxi cerebri were also present together with the saggital venous sinuses. The contents of the posterior fossa were present and normal. The heart, lungs, and abdominal viscera were all normal.

A diagnosis of thalamic failure in hydranencephaly was then made as the cause of death.

Figure II: The postmortem findings demonstrating the absence of the cerebral hemispheres.

Figure III: The postmortem findings demonstrating the absence of the cerebral hemispheres.



Discussion

Hydranencephaly is considered to be the result of a destructive process or lesion in previously normal brain. The insult can be at any time from the 11th week of gestation through the first postnatal year [3]. This results in massive hemispheric necrosis. Debris removal leaves bilateral cystic structures, usually sparing the basal ganglia, the brain stem and the cerebellum.[7]

The typical vascular like distribution of the lesion in hydranencephaly led to the hypothesis of ischaemic etiology partially confirmed by animal models of carotid ligation reproducing the gross appearance of the disease. Involvement of one lobe ;hemihydranencephaly has also been described in the literature.[14,16] The case in discussion involved all two cerebral hemispheres.

Causes

Most of the cases of hydranencephaly are sporadic, with a minority, which is very rare, known as Fowler type hydranencephaly occurring as an autosomal recessive inheritance disease. [6,8,17] The most commonly suspected non-genetic causes of hydranencephaly are in utero vascular accidents, toxic drug use, infections such as toxoplasma gondii, cytomegalovirus, and herpes simplex which are known to cause vasculitis or encephalitis. Also some other viral infections have been identified as causes in animals; for exanple, the akabane and the Aino viruses in cattle.[9,15]

The incidence of hydranencephaly is increased in fetuses of smoking mothers.[3]The cause in this case being was not known.

Hydranencephaly is readily diagnosed in utero per ultrasound scan. In the developing world, hydranencephaly diagnosed postnatally is well documented. [13] This is most likely due to the limited access to fetal ultrasonography. The characteristic clinical picture of presentation has been described.[2]

At birth and for several weeks post natal, unless enlargement of the head becomes noticeable, the behaviour of these infants appear to differ in no respect from normal ones. From the age of 2weeks to 3months, these children tend to develop enlargement of the head. Hyperirritability becomes apparent coincident with enlargement of the head, and is often associated with the Moro, the grasp and the startle reflexes. Incoordinated movements of the eyes, strabimus and nystamus are frequently found.

The occurrence of facial nerve palsy was not found in other articles from a pubmed search. Although the pupils reacted to light, the evidence of true visual appreciation could not be demonstrated. Convulsions which can range from absence attacks to intermittent rigidity and twitching movements of the face and extremities occur.

The cracked pot sign is elicited on percussing the head. Transillumination of the fluid filled cranium is an important sign suggestive of possible hydranencephaly.

For many years, hydranencephaly was suspected on the basis of transillumination and remained a post mortem curosity.[2] The importance of transillumination has decreased with the availability of cross sectional imaging.

Hydranencephaly can now be readily diagnosed postnatally as well in utero by ultrasonography.[3] Magnetic resonance imaging is considered the best modality for the overall evaluation of the anomaly and the documentation of cortical remnants. On computed tomography scan, cortical tissue below the parietal bone convexity may be overlooked with the resultant misdiagnosis of cases of hydrocephalus as hydranencephaly.

The differentials which must be excluded include severe alobar holoprosencephaly and severe hydrocephalus. Treatment given to children with hydranencephaly does not improve the outcome.[13,20]

If the patient's head circumference is increasing, shunting may be needed to stop the development of megalencephaly, for easy of nursing care. [3]

This was the reason why a shunt was inserted. Other treatments given are symptomatic treatments. For example, phenobarbital has been used successfully to control seizures.[5]

The life expectancy of most patients with hydranencephaly is markedly reduced many being either still born or surviving less than a year although there are reports of prolonged survivors. [3,5]

Prolonged survivors can cause significant discomfort to the caregivers and even more so if their heads are allowed to grow two big.

Temperature Control And Hydranencephaly.

The thalamus and hypothalamus are essential structures for temperature control. Significant abnormalities of the brainstem and thalamus have been documented in many, but not all of those infants who die within days to weeks of birth. [5] The presence of the upper motor facial nerve palsy in this case is a sign of microscopic abnormality of the brain stem and or the thalamus despite the normal gross assessment at

postmortem.

It has been postulated that the circuits necessary for maintenance of temperature, blood pressure and cardiorespiratory function are, at least in part, functional in prolonged survivors.[5] Poor temperature control and the presence of upper motor facial nerve palsy could be important parameters whose presence may preclude the insertion of a venticuloperitoneal shunt considering the possible risk of intractable hypothermia.

A currently ongoing debate to modify the current brain death criteria for infants with anencephaly (with hydranencephaly included) so that their organs can be more readily procured for transplantation may perhaps be mentioned in this discussion. The problem of poor temperature control as a poor prognostic sign, if fully investigated, may be a significant contributor to this debate.

Conclusion

Episodes of preoperative poor temperature control especially if associated with upper motor neuron facial nerve palsy may need to be considered as a bad prognostic sign and will require further evaluation as a basis upon which shunting for hydrocephalus may be precluded to avoid table deaths.

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